



Hemepath Case 52: 38-Year-Old Male

HISTORY

A 38-year-old male of Hispanic origin presents with a 3-day history of severe epistaxis. There is no history of trauma and no family history of bleeding disorders. He is not taking any medications or recreational drugs. The patient also complains of general malaise, chills, and drenching night sweats. He had gingival bleeding 2 weeks ago, which stopped after 10 days. On examination, the patient is noted to be feverish.

CBC

Hgb (g/L)	Low
MCV	N
Reticulocyte Count	Low
WBC	Low
Plt	Low

DESCRIPTION OF SLIDE

Peripheral Blood Smear

The peripheral blood smear contains a high percentage of blasts (see circles): they are variable in size and have moderately abundant granular cytoplasm. Many cells contain Auer rods and some cells have multiple Auer rods (see rectangles). Residual normal hematopoiesis is limited and shows no significant dysplasia.

*** To see the slide annotations in Imagescope, click on VIEW, then ANNOTATIONS, and then on the "eye" icon adjacent to the word "Layers". In the "Layer Attributes" box, a brief description of the annotations is provided. You may also click on individual layer region (e.g. region 1) in the "Layer Regions" box to locate each annotation – this is especially helpful in identifying annotations when the slide is not zoomed in. ***

MORPHOLOGICAL DIAGNOSIS

Acute promyelocytic leukemia

DISCUSSION

Acute promyelocytic leukemia (APL) is the M3 subtype of acute myelogenous leukemia (AML) under the French-American-British (FAB) classification. In the WHO system it is now called AML with translocation 15;17. Classically, APL is characterized by blasts containing numerous Auer rods. Most patients have the t(15;17), creating the fusion genes PML-RAR α (promyelocytic leukemia-retinoic acid receptor α) and RAR α -PML on der(15) and der(17) respectively. The PML-RAR α protein acts as an abnormal retinoid

receptor with malignant potential. There are also examples of other mutations presenting with APL, but these are rare.

Unlike other subtypes of AML, APL has a propensity to affect younger patients, with a clinical picture of pancytopenia rather than leukocytosis. In fact, leukocytosis in APL patients is a poor prognostic factor. Patients often present with fatigue and constitutional symptoms, as well as severe hemorrhage. Disseminated intravascular coagulopathy (DIC) may also occur in APL owing to activation of the coagulation system by abnormal granules released from malignant promyelocytes, and is considered a medical emergency.

One variant of APL, the hypogranular or microgranular variant, may be somewhat more challenging to diagnose because the classic hypergranular morphology is lacking. These cases may require flow cytometry and/or cytogenetics to confirm a diagnosis.